BSR and BHPR guidelines for the management of giant cell arteritis

Bhaskar Dasgupta1, Frances A. Borg1, Nada Hassan1, Leslie Alexander1, Kevin Barraclough2, Brian Bourke3, Joan Fulcher4, Jane Hollywood1, Andrew Hutchings5, Pat James4, Valerie Kyle6, Jennifer Nott7, Michael Power8 and Ash Samanta9 on behalf of the BSR and BHPR Standards, Guidelines and Audit Working Group

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Executive summary

Scope and purpose

GCA is the commonest of all the vasculitides. Visual loss occurs in up to one-fifth of patients, which may be preventable by prompt recognition and treatment [1,2].

The aim of these guidelines is to encourage the prompt diagnosis and management of GCA, with emphasis on the prevention of visual loss. Their scope is to provide evidence-based advice for the assessment and diagnosis of GCA, for initial and further management and for monitoring of disease activity, complications and relapse. This is a summary of the guidelines and the full guideline is available at Rheumatology online.

The guidelines

The recommendations for the guidelines are set out in points 1 to 9.

(1) Early recognition and diagnosis of GCA is paramount [2]. Particular attention should be paid to the predictive features of ischaemic neuro-ophthalmic complications (C).

A patient >50 years of age presenting with the following features should raise suspicion of GCA:

- Abrupt-onset headache (usually unilateral in the temporal area).
- Scalp tenderness.
- Jaw and tongue claudication.
- Visual symptoms (including diplopia).
- Constitutional symptoms.
- Polymyalgic symptoms.
- Limb claudication.

Examination may show:

- Abnormal superficial temporal artery: tender, thickened with reduced or absent pulsation.
- Scalp tenderness.
- Transient or permanent visual loss.
- Visual field defect.
- Relative afferent papillary defect.
- Anterior ischaemic optic neuritis.
- Central retinal artery occlusion.

1Department of Rheumatology, Southend University Hospital, Essex.
2Painswick Centre, Painswick.
3Department of Rheumatology, St George’s Hospital, London.
4PMRGCA Group, Southend & Essex.
5Health Services Research, London School of Hygiene and Tropical Medicine, London.
6Department of Rheumatology, Frenchay Hospital, Bristol.
7East Anglia PMR and GCA Support Group, Ipswich.
8Clinical Knowledge Summaries Service, Sowerby Health Informatics, Newcastle upon Tyne.
9Leicester Royal Infirmary, Leicester, UK.


Correspondence to: Bhaskar Dasgupta, Department of Rheumatology, Southend University Hospital, Prittlewell Chase, Westcliff-on-Sea, Essex SS0 0RY, UK. E-mail: bhaskar.dasgupta@southend.nhs.uk
● Upper cranial nerve palsies.
● Features of large-vessel GCA: vascular bruits and asymmetry of pulses or blood pressure.

Laboratory investigations include:
● Full blood count, urea and electrolytes, liver function tests, CRP, ESR.
● An acute-phase response is the characteristic of GCA (raised ESR, CRP, anaemia, thrombocytosis, abnormal liver function tests, particularly raised alkaline phosphatase, raised α1 and α2 globulins on serum electrophoresis). However, GCA can occur in the face of lower levels of inflammatory markers, if the clinical picture is typical.
● Chest radiograph.
● Urinalysis.
● Other relevant investigations to exclude mimicking conditions.

Features predictive of ischaemic neuro-ophthalmic complications [3, 4]:
● Jaw claudication.
● Diplopia.
● Temporal artery abnormalities.

(2) Urgent referral for specialist evaluation is suggested for all patients with GCA. Temporal artery biopsy (TAB) should be considered whenever a diagnosis of GCA is suspected. This should not delay the prompt institution of high-dose glucocorticosteroid therapy (C).

TAB can remain positive for 2–6 weeks after the commencement of treatment. It should be performed by a surgical unit experienced in regular TAB, and samples should be at least 1 cm in length. Contralateral biopsy is usually unnecessary.

TAB may be negative in some patients. They should be regarded as having GCA if there is a typical clinical picture and response to glucocorticosteroids.

(3) Imaging techniques show promise for the diagnosis and monitoring of GCA. However, these do not replace TAB for cranial GCA. Their role in early diagnosis of cranial GCA is an important area of future research (B).

The use of duplex ultrasound is currently limited as it requires a high level of experience and training. Other imaging modalities (PET and MRI) should be currently reserved for investigation of suspected large-vessel GCA.

(4a) High-dose glucocorticosteroid therapy should be initiated immediately when clinical suspicion of GCA is raised (C).

Recommended starting dosages of glucocorticosteroids are:
● Uncomplicated GCA (no jaw claudication or visual disturbance): 40–60 mg prednisolone daily.
● Evolving visual loss or amaurosis fugax (complicated GCA): 500 mg to 1 g of i.v. methylprednisolone for 3 days before oral glucocorticosteroids.
● Established visual loss: 60 mg prednisolone daily to protect the contralateral eye.

Patients should also receive bone protection. Proton pump inhibitors for gastrointestinal protection should be considered.

The symptoms of GCA should respond rapidly to high-dose glucocorticosteroid treatment, followed by resolution of the inflammatory response. Failure to do so should raise the question of an alternative diagnosis.

(4b) Glucocorticosteroid reduction should be considered only in the absence of clinical symptoms, signs and laboratory abnormalities suggestive of active disease (C).

This should be balanced against the need to use the lowest effective dose, patient wishes and glucocorticosteroid side effects. Steroid reduction may also be appropriate if the acute-phase response is deemed to be due to another cause.

Suggested tapering regimen:
● 40–60 mg prednisolone continued until symptoms and laboratory abnormalities resolve (at least 3–4 weeks);
● then dose is reduced by 10 mg every 2 weeks to 20 mg;
● then by 2.5 mg every 2–4 weeks to 10 mg; and
● then by 1 mg every 1–2 months provided there is no relapse.

The dose may need adjustment for disease severity, comorbid factors, fracture risk, patient wishes and adverse events. There are also some patients who will require long-term low-dose glucocorticosteroid therapy.

(5) Low-dose aspirin should be considered in patients with GCA if no contraindications exist (C).

(6) Large-vessel GCA should be suspected in patients with prominent systemic symptoms, limb claudication or persistently high-inflammatory markers despite adequate glucocorticosteroid therapy. Imaging techniques, such as PET and MRI scanning, should be reserved for the assessment of suspected large-vessel involvement [5] (C).

(7) Monitoring of therapy should be clinical and supported by the measurement of inflammatory markers (C; this is a consensus statement).

Patients should be monitored for evidence of relapse, disease-related complications and glucocorticosteroid-related complications.

In particular, the following features should be sought:
● Jaw and tongue claudication.
● Visual symptoms.
● Vascular claudication of limbs, bruits and asymmetrical pulses.
● Polymyalgic symptoms.
● Osteoporotic risk factors and fractures.
- Other glucocorticosteroid-related complications.
- Other symptoms that may suggest an alternative diagnosis.

The following investigations should be performed:
- At each visit: full blood count, ESR/CRP, urea and electrolytes, glucose.
- Every 2 years: chest radiograph to monitor for aortic aneurysm (echocardiography, PET and MRI may also be appropriate).
- Bone mineral density may be required.

Routine follow-up should be planned at:
- Weeks 0, 1, 3, 6, then Months 3, 6, 9, 12 in the first year.
- Later (Month 3 onwards) follow-up can be undertaken under shared care.

Relapse:
- Disease relapse should be suspected in patients with a return of symptoms of GCA, ischaemic complications, unexplained fever or polymyalgic symptoms.

**Fig. 1** Approach to diagnosis and management of GCA.

**Key features**
- Abrupt new headache
- Scalp pain and tenderness
- Jaw claudication
- Visual symptoms, e.g. diplopia
- Symptoms of PMR
- Temporal artery abnormalities
- Raised ESR/CRP

**Immediate start of glucocorticosteroid therapy**
- Uncomplicated: without jaw claudication or visual symptoms
  - Prednisolone 40 mg daily
- Complicated: jaw claudication or visual symptoms
  - Prednisolone 60 mg daily

**Urgent referral for specialist management**
- TAB
- Ophthalmological assessment (with ischaemic features)

**Biopsy positive**
- Bone protection

**Biopsy negative**
- Specialist review
  - Clinical suspicion high or US suggests GCA or complications typical of GCA (e.g. anterior ischaemic optic neuritis)
  - Treat as biopsy-positive GCA
- Specialist review
  - Clinical suspicion low
  - Features considered atypical or alternative explanations available
  - Rapid glucocorticosteroid tapering (within 2 weeks)
  - Treat alternative diagnosis

**Gradual glucocorticosteroid tapering after disease control**
- Monitoring:
  - Disease activity related:
    - relapses, large-vessel GCA
  - Treatment related:
    - weight, fractures, blood pressure, glucose, cataracts, glaucoma, lipids, skin
  - Consider MTX

**Early recognition of GCA**
- Irreversible ischaemic complications, such as vision loss, occur almost always early, prior to glucocorticosteroid therapy

**References**

- Abrupt new headache
- Scalp pain and tenderness
- Jaw claudication
- Visual symptoms, e.g. diplopia
- Symptoms of PMR
- Temporal artery abnormalities
- Raised ESR/CRP
A rise in ESR/CRP is usually seen with relapse, but relapse can be seen with normal inflammatory markers.

- All patients in whom relapse is suspected should be treated as below, and discussed or referred for specialist assessment.
- Return of headache should be treated with the previous higher dose of glucocorticosteroids.
- Jaw claudication requires 60 mg prednisolone.
- Eye symptoms need the use of either 60 mg prednisolone or i.v. methylprednisolone.
- Symptoms of large-vessel disease should prompt further investigation with MRI or PET and the use of systemic vasculitis treatment protocols.

For recurrent relapse, see below.

(8) The early introduction of MTX or alternative immunosuppressants should be considered as adjuvant therapy (B).

Recurrent relapse or failure to wean glucocorticosteroid dose requires the consideration of adjuvant therapy, such as MTX or other immunosuppressants. These immunosuppressive agents should be started at the third relapse. Biological therapies still require further study, and are not yet recommended.

(9) Patient education (D).

We suggest developing a new Arthritis Research Campaign booklet on GCA for the use of newly diagnosed patients. The approach to diagnosis and management of GCA is summarized in Figure 1.

Recommendations for audit

Audit standards should include the minimum baseline data set recorded, initial glucocorticosteroid dose and taper, monitoring frequency and outcomes. The key performance measure should be the time from symptoms to initial treatment.

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